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The Case | Thirty-one-year old woman with hypertension and abnormal renal imaging

K Kiryluk[1], RA Rabenou[2], ER Goldberg[3] and M Gupta[4]

[1]Division of Nephrology, Department of Medicine, Columbia University, College of Physicians and Surgeons, New York, New York, USA; [2]Division of Nephrology, Department of Medicine, New York University School of Medicine, New York, New York, USA; [3]Division of General Internal Medicine, Department of Medicine, New York University School of Medicine, New York, New York, USA and [4]Department of Urology, Columbia University, College of Physicians and Surgeons, New York, New York, USA

Correspondence: *K Kiryluk, Division of Nephrology, College of Physicians and Surgeons, Columbia University, 622 West 168th Street, PH4 Stem – Room 124, New York, New York 10032, USA.*
E-mail: kk473@columbia.edu

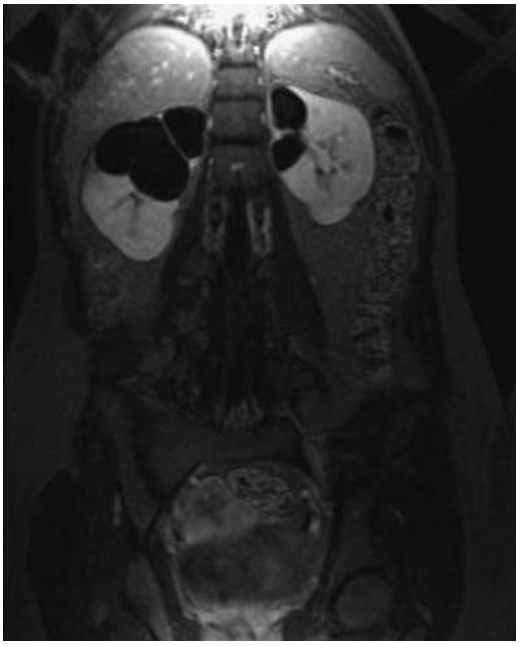

Figure 1 | Standard magnetic resonance imaging.

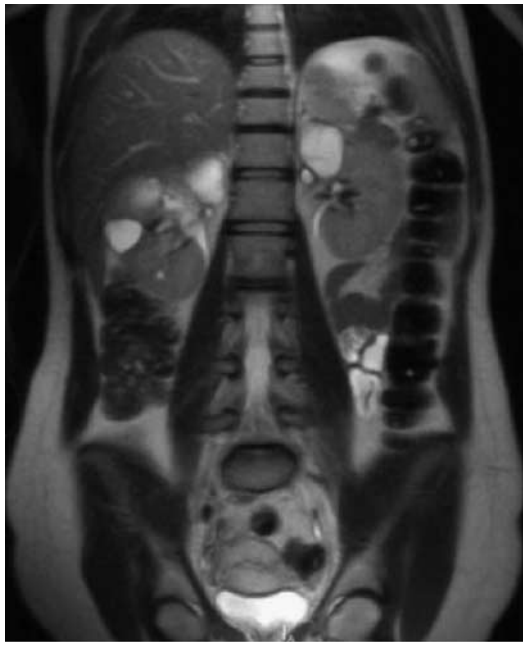

Figure 2 | Magnetic resonance urography.

A 31-year-old woman presented with headaches and high blood pressure (180/100 mm Hg). She had a history of four kidney infections in the last 10 years. There was no family history of kidney disease. Her physical exam was unremarkable, the creatinine was 1.0 mg per 100 ml (Ref. 0.5–0.9), blood urea nitrogen 17 mg per 100 ml (Ref. 7–20), and urinalysis was normal. Renal ultrasonography revealed three large cysts in the right kidney (2.8 cm upper pole, 4.4 cm mid portion, and 2.4 cm lower pole) and two cysts in the left kidney (4.1 and 1.9 cm, both at the upper pole). Doppler evaluation was negative for renal artery stenoses. Magnetic resonance imaging was performed (Figures 1 and 2).

What is the diagnosis?

SEE NEXT PAGE FOR ANSWERS

The Diagnosis | Bilateral infundibular stenoses

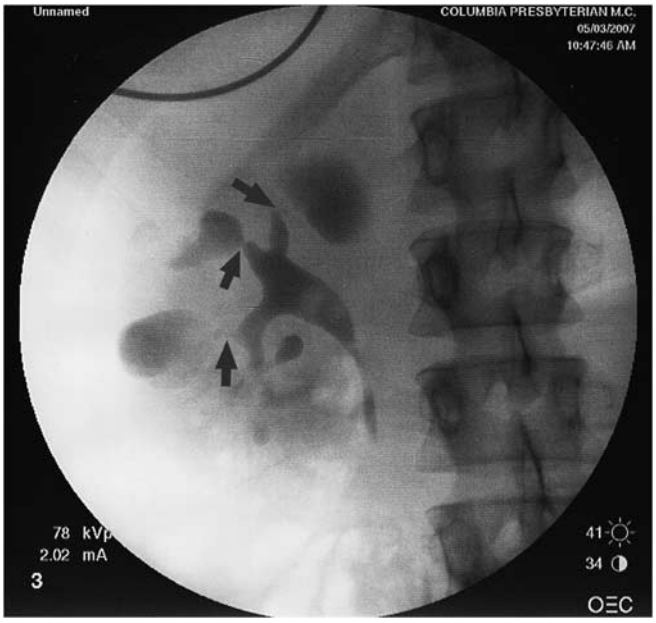

Figure 3 | Retrograde ureteropyelogram of the right kidney. The architecture of the renal collecting system is distorted with evidence of three massive hydrocalices and infundibular stenoses (arrows).

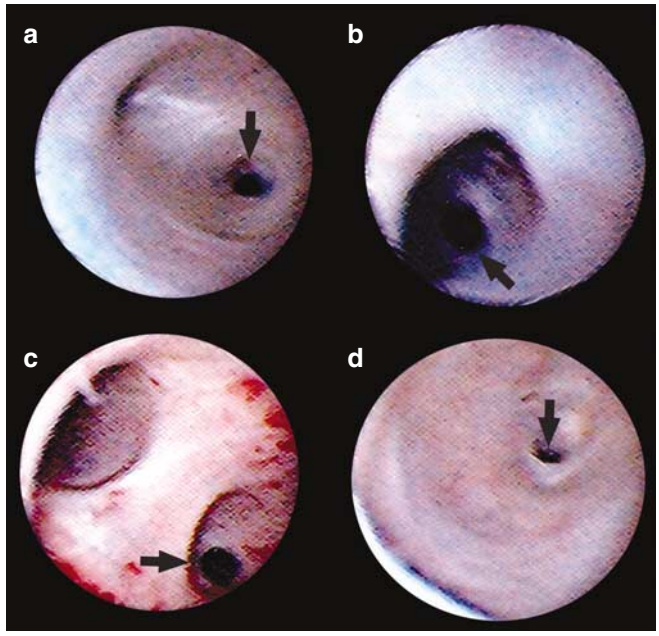

Figure 4 | Endoscopic images of the stenotic infundibula (arrows). Retrograde view from the renal pelvis directly visualizing stenotic infundibula (**a–d**) that were too narrow to accomodate anything but a small (365 μm) laser fiber.

In this case, simultaneous presence of contrast within the 'cysts', renal pelvices, ureters, and bladder detected on MR urography (Figure 2) was suggestive of a collecting system abnormality. However, retrograde ureteropyelogram was required to establish a definitive diagnosis (Figure 3). Bilateral infundibular stenoses were then confirmed by nephroscopy (Figure 4). Infundibular stenosis is an obstructive disorder of the intrarenal collecting system that may manifest with hypertension and progressive renal dysfunction. Bilateral stenoses are extremely rare, and can be mistaken for multicystic kidney disease on ultrasound, standard magnetic resonance imaging, or computed tomography. This disorder must be differentiated from polycystic kidney disease, simple renal cysts, caliceal diverticula, and megacalicosis (idiopathic caliceal dilation without infundibular narrowing).

Infundibular stenosis may be congenital or acquired secondary to infection, tumor, or trauma. Hydrocalices arise when renal calices are obstructed by narrowed infundibula. Bilateral cases are most often congenital. Infundibulopelvic anomalies may represent a link in a clinical spectrum of obstructed dysmorphic kidneys that extends from multicystic renal dysplasia to hydronephrosis.[1] Several other urogenital malformations have been observed in association with infundibular stenoses. These include malrotated kidney, megaureter, renal agenesis, renal dysplasia, and vesicoureteral reflux.[2] Although most described cases are sporadic, congenital infundibulopelvic anomalies have been reported in families with other kidney malformations, such as renal hypodysplasia.[3] This observation suggests that genetic factors may be involved in the pathogenesis of infundibular stenosis.

Patients with congenital hydrocalices can develop hypertension and renal insufficiency and progress to end-stage renal disease. Thus, symptomatic infundibular stenosis should be treated promptly. Treatment involves decompression of hydrocalices and relief of obstruction, both of which can be successfully accomplished by minimally invasive flexible ureteroscopy.[4] In this case, a Holmium-YAG laser was used to perform infundibuloplasties and drainage of right-sided hydrocalices. Post-procedure imaging demonstrated significant reduction of hydrocalycosis on the right side and the patient is scheduled to return for a left-sided procedure.

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